REPORT OF A CASE OF ANENCEPHALY, WITH A MICROSCOPICAL STUDY BEARING ON ITS RELATION TO THE SENSORY AND MOTOR TRACTS.

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CASES in which infants are born at full term with no fore-brain are rare. Syme describes a case, *Edinb. Med. and Surg. Journal*, vol. xxiv, p. 295, in which the infant lived six months. After death only the cerebellum, pons, and parts below were found. Panizza, of Paris, reports a similar case that lived eighteen hours (Gintrac, Maladies de l'appareil nerv., Paris, 1867, v. i., p. 51). Ollivier reports one that lived twenty hours (Maladies de la moelle epiniere, Paris, 1837, v. i., p. 179). The two cases of Rohon and of Starr have been reported recently, and are familiar.

This form of monstrosity, so far as I can judge, would be called by Geoffry St. Hillaire "pseudo-encephaly."

The interest in such cases previous to the discovery of the modern methods of studying nerve anatomy has been purely clinical and teratological.

We know now, however, that all such cases of congenital anomalies of the nervous system constitute natural experiments in the atrophy method, and that one of the chief

interests in monsters lies in what they tell us post-mortem regarding nerve tracts and centres.

CLINICAL HISTORY.—The case is that of a male infant born at full term. The mother was unmarried, but was healthy, denied syphilis or anything unusual in the mode of sexual intercourse or incidents of pregnancy. She had had no other child. The father, she said, was healthy.

The child was large for its age, must have weighed eight or nine pounds, was very plump and healthy-looking. It showed no deformity except that the head was peculiarly shaped. It was large proportionately, the forehead narrow, and the whole head very narrow and long in the occipital-frontal diameter. The sutures were not united, and the bones were freely movable. The eyes were generally closed, but occasionally opened. The child nursed a little, but had difficulty in swallowing, and after feeding would have attacks of cyanosis.

It was noticed that it cried very little, and only in a feeble way. It had natural movements of the bowels and bladder. It had had no convulsions or rigidity or paralysis. On pinching or pushing it, the infant cried. It lived two and one-half days.

AUTOPSY.—On puncturing the membrane between the cranial sutures, a yellowish liquid spurted out. Cutting a way the calvarium, the whole cranial cavity above the tentorium was found perfectly empty and smooth. The anterior and middle fossæ contained nothing but the foldings of the membranes. In the posterior fossa the tentorium was bare, but the cerebellum could be felt beneath it.

The cerebellum was fully developed and of good size, measuring 6½ ctm. in width and 3½ ctm. anterio-posteriorly. The roots of all the cranial nerves were present except the first. The optic nerves were small, and the chiasm scarcely to be made out. The optic tracts showed barely a trace. The third nerves were well developed. The fifth nerves were all well developed, and both motor and sensory roots could be seen. They arose far out on the lateral surface of the pons in the sulcus between it and the cerebellum. The

other cranial nerves could be made out in the sulcus external to the olives, the hypoglossal apparently arising from the lower outer border of the olives.

The vertebral arteries joined to form the basilar, which at the cephalic end of the pons abruptly terminated in four small arteries. The two anterior of these passed forward to the optic thalami, the two posterior to the cerebellum.

The pons varolii was very small. The medulla appeared to be made up mostly of the two large olivary bodies, which almost met in the median line. The corpora quadrigemina seemed fairly well developed. Anterior and external to these were two lobes, evidently parts of the optic thalamus, and the sole representation of the first cerebral vesicle. The left was much the larger, and measured, in a direction from outside and posterior up and in, 3 ctm. The spinal cord appeared normal, but small.

The specimen was hardened in Muller's fluid and then in alcohol. Sections were made at the level of the third to fourth lumbar nerves, eighth to tenth and fourth to sixth dorsal, fourth to sixth and first cervical.

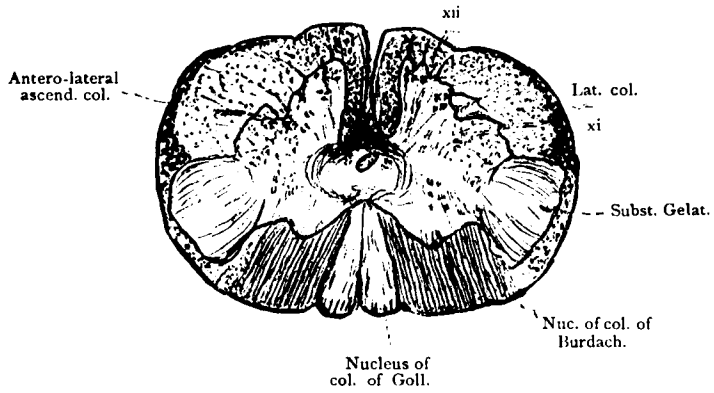
Sections were also made, at different levels, through the medulla and pons. These sections were stained in Weigert's and ordinary hæmatoxylon, carmine, and aniline blue.

The most interest attaches to the appearances in the pons and medulla, and these will be described first. The cerebral hemispheres and corpora striata being entirely absent, as already stated, we should expect to find absent all those uninterrupted tracts depending for their nutrition upon the integrity of the parts mentioned. The afferent tracts to the brain should be present.

MICROSCOPICAL APPEARANCES.—Section I. Beginning from below, I find, in sections at the level of the motor decussation, absence almost entirely of this decussion and of the lateral pyramidal tracts. In place of this, one sees only a decussation of sensory fibres starting from the nuclei of the post-median and post-external columns. The fibres cross over at the bottom of the anterior median fissure, part (1)

turn up directly, and part (2) pass anteriorly along the median surface of the anterior fissure to go towards the olivary body. The first set (1) form the ascending fibres of the inter-olivary tract.

Posterior to the sensory decussation is a decussation of a few fibres connecting the cells of the anterior horns.



The post-median and post-external nuclei are well developed, as are also the corresponding columns at a lower level.

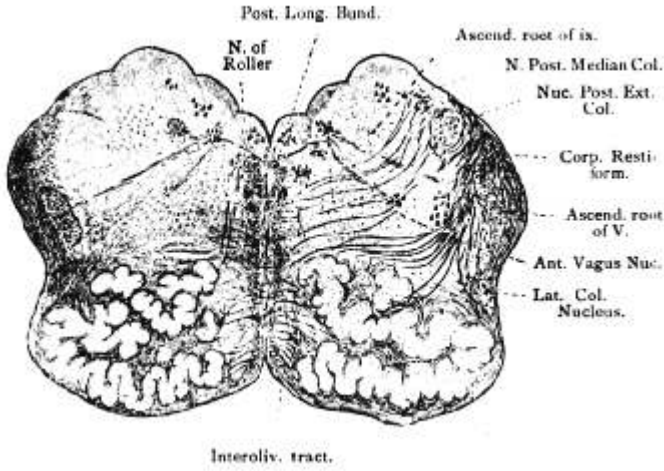
The gray matter is normal in arrangement and in its cells, though the anterior horns are a little smaller than normal. The place of the crossed pyramidal tracts is occupied only by a few nerve fibres.

The anterior columns are abnormally narrow. The direct cerebellar tracts are well and distinctly marked. There is apparently only a partial development of the antero lateral ascending column.

The anterior and posterior nerve roots are well developed. The fibres of origin of the ascending root of the fifth are visible. The nuclei and fibres of origin of the eleventh and twelfth nerves can be seen. Fibres can also be seen to pass from the posterior columns into the lateral column and lateral part of intermediate gray, then turning up to enter at a higher level, the formatio-reticularis.

SECTION II.—In the next series of sections, at the level of the lower part of the olive, the pyramids are seen to be entirely absent, so that the olives form the ventral edge of the section.

The interolivary tract is present, but is very poorly developed. The part best seen is the ventral portion lying almost at the bottom of the fissure between the olives. This ventral portion of the interolivary tract is seen to be made

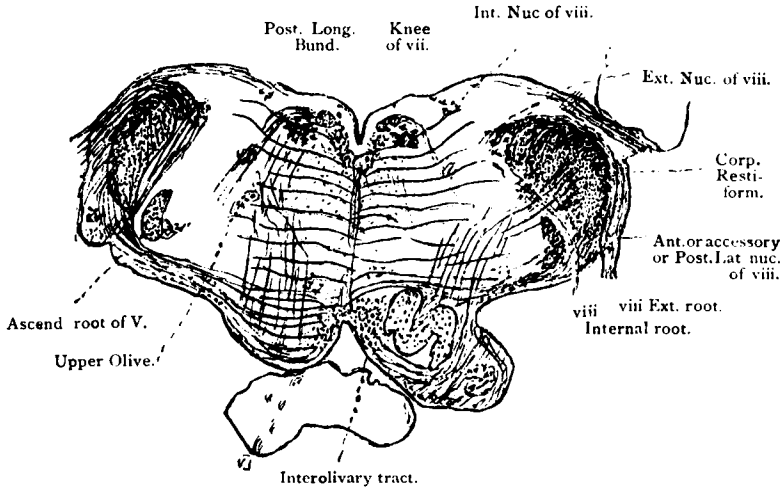


up from internal and anterior arcuati fibres from the post-median and post-external column nuclei. Posteriorly, the longitudinal bundle is seen normal in size. The nucleus of Roller is present. The formatio-reticularis is of nearly normal appearance, but not quite so rich in fibres as it should be ; the internal and external olives are present.

The respiratory bundle is normal. The anterio-posterior fibres of the raphe are numerous, but seem to end abruptly at the bottom of the anterior median fissure between the olives. The raphe fibres in general are much more numerous in the interolivary portion, as is the case in normal cords.

The lateral column fibres and nucleus are very distinct. The gray matter and cranial nuclei on the floor of the ventricle are normal.

SECTION III.—The next section is made at the upper border of the olives and lower edge of the pons. The interolivary tract is quite distinct in the ventral portion between the olives. Its middle and dorsal portions are fairly represented, while the posterior longitudinal bundle is very distinct. The anterior arcuate fibres pass in a distinct strand



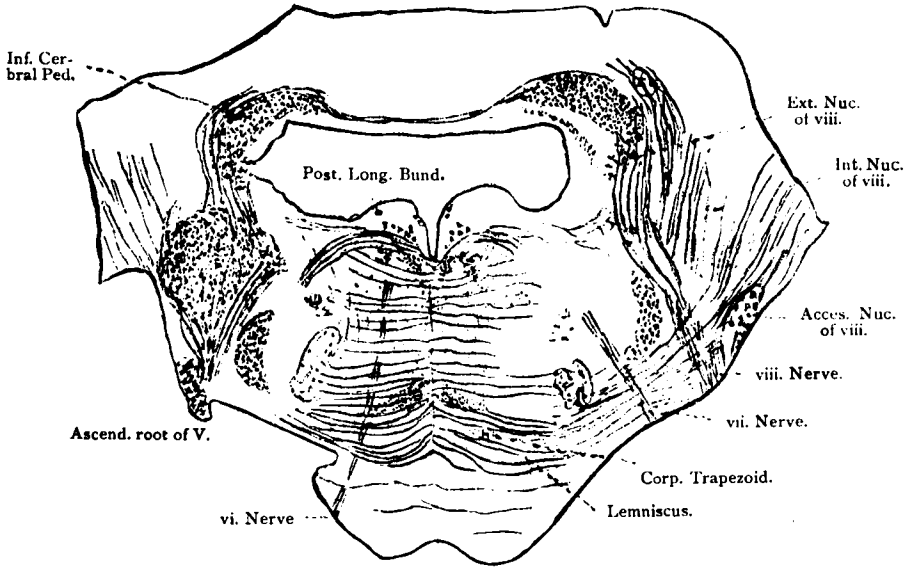
from the corpus restiforme to the ventral part of the raphe. Here some cross and some enter the lemniscus. The fibres of the raphe are fully developed dorsally.

The nuclei of Roller, which is supposed by Bechterew to be the end station of the fibres of the lateral fundamental column, are here very distinct. The formatio-reticularis is rather poor in fibres. What corresponds to the caudal edge of the pons is an an irregular mass of undifferentiated tissue through which the sixth cranial nerves run, and in which can be seen a few fully developed transverse nerve fibres.

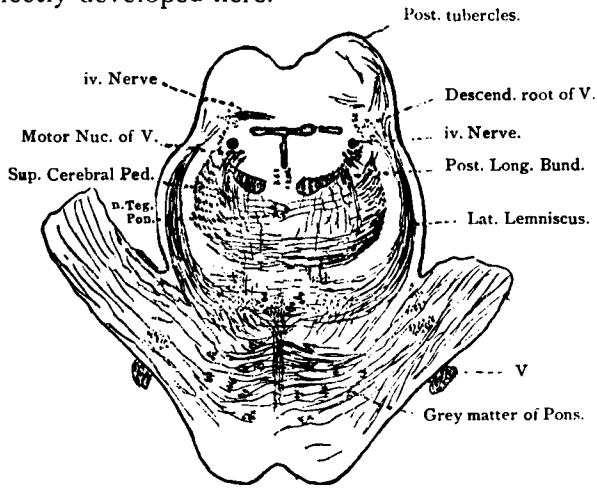
The dorso-ventral or linear fibres of the raphe are moderately developed, and are most numerous ventrally. Here they come *via* the anterior arcuate fibres from the corpus restiforme ; they pass up, and having crossed, enter the lemniscus.

It thus appears that the fibres of the raphe serve to connect the cranial nerve nuclei with the pyramids, and also with the lemniscus or corpus restiforme.

SECTION IV.—At a higher level of the pons, but still below the middle and at about the level of the nucleus of the sixth, the lemniscus is seen in its altered position lying



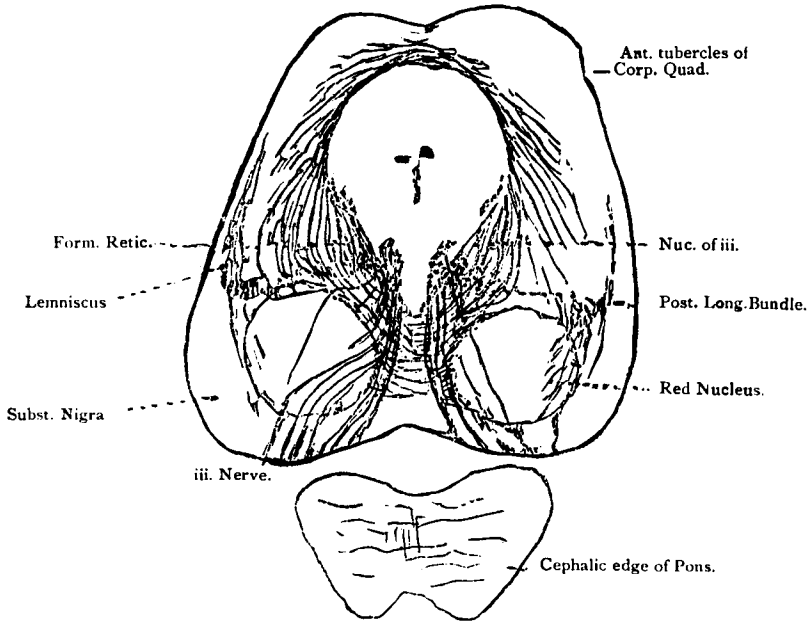
horizontally upon the fibres of the corpus trapezoid, which are here very well marked. It is here small. The fibres of the raphe are very few. The formatio-reticularis is but imperfectly developed here.



SECTION V.—In a section still higher, through the upper part of the pons and the posterior tubercles of the corpora

quadrigemina, there is a very distinct development of the lateral or lower lemniscus, while the median or upper lemniscus is small.

SECTION VI.—At a still higher level, just through the red nuclei and touching the cephalic edge of the pons, one notes the entire absence of cerebral peduncles. The red nuclei are apparently well developed.



The median lemniscus is now plainly visible, also the nuclei and fibres of the third nerve. The lateral lemniscus has ended in the posterior tubercles, and is not seen. Fibres can be seen coming from the posterior commissure and passing down to enter the posterior longitudinal bundle. There is no lenticular loop. The corpora quadrigemina seem small and almost structureless, save for the fibres of the posterior commissure which run through its lower part. There is still some evidence of the formatio-reticularis. The substantia nigra is present, but imperfectly developed.

At this level, the dorsal part of the raphe contains no fibres, the lower or ventral part is very distinct, The fibre

bundle from the inner part of crusta is absent. The fibre bundle from the posterior commissure is absent. The offshoot bundle of Wernicke is absent. The nucleus reticularis-tegmenti-pontis sends down fibres through the raphe. This nucleus, according to Bechterew, is the end station of fibres of anterior fundamental column.

The aqueduct of Sylvius is imperfect. The posterior longitudinal bundle is well marked, as are also the fibres from the anterior cerebellar peduncle. The descending root of the fifth is here visible. The nuclei of the pons are present.

Sections through the optic thalami showed a nearly structureless mass containing but a few nerve fibres and cells.

SUMMARY: The pyramidal tracts are absent, and in consequence the shape and relations of the parts are changed.

The formatio reticularis is apparently normal.

The cranial nerves and nuclei, the respiratory bundle, posterior longitudinal bundle, olives and supplementary olives, and in inner and outer nuclei of Roller, are present and normal.

The sensory decussation and interolivary tract and lemiscus are present, but the median portion is about half the normal size.

In the lower sections the ventral portion of the interolivary tract alone is normal in size, the parts dorsal to it being barely visible; at higher levels (upper olive), the tract in question lies as it should in the fibres of the corpus trapezoid, but is small.

At still higher levels (crossing of the anterior cerebellum peduncles) the lateral or lower lemniscus is very clearly defined, the median very scanty in fibres. At the level of the red nuclei the median part is quite distinct.

The following, therefore, is the condition of the lemniscal tracts:

The innermost bundle to the crusta is not present.

The median lemniscus is about half the normal size, but is traceable as far as the anterior tubercles of the corpora quadrigemina, which it appears to enter. This (the median) is the lemniscal tract which goes partly to the anterior tubercles, partly to the optic thalamus or Luys' body, partly to the lenticular nucleus and parietal cortex (*Hauptschleife*, Bechterew, *Rindenschleife* of Monakow, etc., upper lemniscus.

In experimental atrophies the degeneration has usually been downwards, but in Meyer's case it was upwards *in toto*, and it evidently consists, in large part at least, of afferent fibres.

The lateral lemniscus (lower), which begins chiefly in the upper olive, by which it is connected with the posterior branch of the acoustic nerve (Bechterew, Flechsig, Obersteiner), is present, and apparently ends in the posterior tubercles. This tract is doubtless, therefore, afferent.

The tegmental part only of the crura cerebri is present: The substantia nigra is present, but is structurally but little developed.

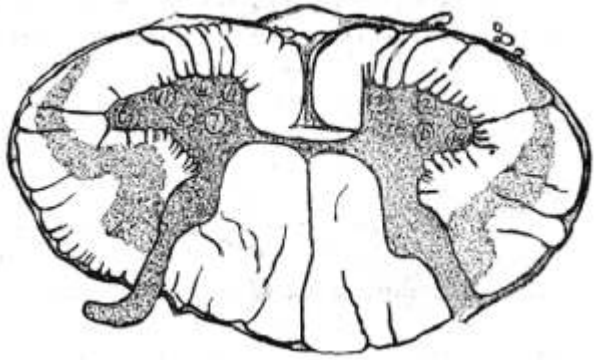
The gray nuclei of the pons are present.

THE SPINAL CORD.—As a whole the cord is smaller than normal.

The diminished size is due to the small anterior and lateral columns. In comparison, the posterior columns look unusually large.

The nerve roots are normal. The anterior columns are much narrower than they should be, but show nothing abnormal otherwise.

The lateral columns reveal a tract of connective or undifferentiated tissue extending throughout the length of the

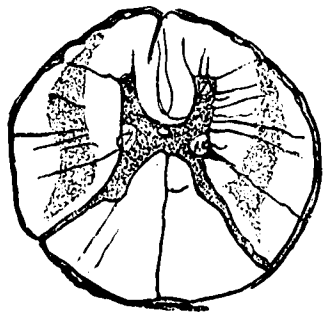


cord. The position of this undeveloped tract in the upper cervical region is peculiar. As shown in the cut, it appears to extend laterally and ventrally to the surface, occupying, in a measure, the region of the anterior ascending tract. A few nerve fibres are present in this region; on transverse

sections of the cord they are cut off obliquely and appear as if running forwards and inwards. The direct cerebellar tract, Lissauer's bundle, which may be called the posterior root column, the postero-external and postero-median columns are normal.

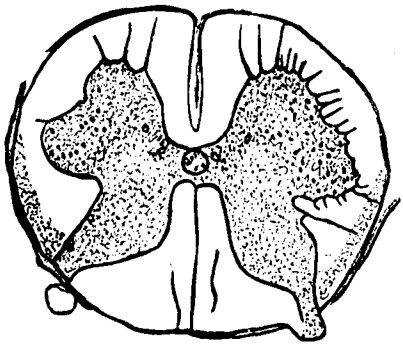
The longitudinal sections were made at the level of the fourth to sixth dorsal roots, the plane of the sections being directed anteriorly, the appearance of the anterior commissure was studied and compared with similar sections in a normal cord.

In the *anencephalic cord the commissure was poorer in fibres, and these crossed directly* from one side to the other at a very acute angle. In the normal cord some fibres crossed in this way, but others ran along the edge of the fissure for a short distance and then cross at a more obtuse angle. This *must represent the mode of crossing* of the direct *pyramidal tract fibres.*



The posterior roots can be seen to enter in two bundles of fibres; one of fine fibres lies external, appears to connect directly with the posterior nerve and Lissauer's bundle. The other divides into fibres which pass through (?) and around inner side of subs. gel. to posterior cornual cells and Clark's cells. These fibres can be traced to the anterior cornu of same side and through the anterior commissure to the anterior cornu of the other side.

The anterior commissure is divided into two parts, one lying anterior to the other, and it is through the posterior one that the posterior cornual fibres pass.



The gray matter of the cord and its cells are fairly developed.